Oral lichen sclerosus: a rare case and update

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Abstract

Lichen sclerosus (LS) is a chronic inflammatory mucocutaneous disease that often affects the anogenital area; oral mucosal lesions are extremely rare. A 52-year-old woman presented for evaluation of an 8-year history of a persistent whitish plaque in the buccal mucosa. Intraoral examination revealed multiple elevated whitish plaques diffusely distributed in the buccal mucosa associated with an area of tissue atrophy. Although both leukoplakia and lichen planus were considered, incisional biopsy and later, full excision confirmed the histopathological diagnosis of oral LS. After 6 months of follow-up, there are no clinical signs of relapse. This case highlights the importance of clinical and histopathological findings for the correct diagnosis and treatment of oral LS.

Keywords: oral lichen sclerosus, oral cavity, oral mucosa

Introduction

Lichen sclerosus (LS) is a chronic inflammatory mucocutaneous disease mainly affecting the skin and anogenital region; oral mucosa lesions appear in only rare cases, mainly as a single clinical manifestation [1-4]. The condition can affect individuals at any age, although it is five-to-ten times more common in females [2]. LS displays the potential to cause functional impairment, as well as, in rare situations, evolve to malignancy [5-6]. Its etiology remains uncertain, although several probable predisposing factors have been reported, such as trauma, genetic susceptibility, hormonal factors, infections, and autoimmunity [2-3].

Clinically, LS oral lesions present as whitish plaques, which may or may not be slightly elevated, ranging from a small localized macule to a lesion involving large mucosal areas [1, 7]. Lichen sclerosus exhibits a stratified squamous epithelium with focal hyperparakeratosis and atrophic areas. The presence of a subepithelial cleft is also noted, with homogenization and hyalinization related to loss of collagen fibers. In addition, one observes the presence of a mononuclear inflammatory infiltrate band, usually located in the deeper portions of the lamina propria underlying the hyalinization zone [1, 2, 4]. In this context, the present report discusses a case of oral lichen sclerosus and assesses clinical and histopathological characteristics in comparison to cases in the literature.

Case Synopsis

A 52-year-old woman sought out the Federal University of Rio Grande do Norte Stomatology Clinic, located in the city of Natal, Rio Grande do Norte, Brazil, complaining of a persistent white spot located in the buccal mucosa. The patient reported that the spot grew slowly over about 8 years, but noted no pain or other discomfort. On intraoral physical examination, multiple slightly elevated whitish plaques, with a fibrous consistency were observed. They were located diffusely in the right buccal mucosa extending to the proximity of the labial commissure and upper vestibule fundus. The largest lesions measured approximately 1.0 x 2.0 cm and the smallest was approximately 0.5cm in diameter. In addition, an area of tissue atrophy was observed adjacent to the clinical lesion, close to the labial commissure (**Figure 1**).

The patient reported no alcoholism or smoking, no history of trauma in the lesion location, and no allergies. Based on the clinical examination, a diagnosis of leukoplakia diagnosis was entertained and an incisional biopsy was performed for confirmation. A histopathological examination revealed the presence of an atrophied hyperparakeratinized stratified squamous epithelium, with areas of exocytosis and a subepithelial cleft (**Figure 2**A). The underlying lamina propria exhibited an intense band of hyalinization (**Figure 2**B), which had in its deeper portion, an intense lymphocytic inflammatory infiltrate within the band (**Figure 2**C). Based on these clinical and histopathological aspects, a diagnosis of oral LS was reached.

The patient was referred to a dermatologist and gynecologist to assess skin and genital lesions, respectively. However, no vulvar or cutaneous LS was found. Thus, total surgical removal of the lesion was performed and the previous histopathological diagnosis of LS was confirmed. The patient was observed periodically, and after 6 months of follow-up, no clinical signs of relapse have been noted (**Figure 3**).

Case Discussion

Lichen sclerosus was first described in 1887 by Hallopeau as a pathological process that affected the anogenital region and was named "atrophic lichen planus." Later, in 1892, Darier studied an injury that exhibited similar histopathological characteristics to the one described by Hallopeau and termed it "sclerosus lichen planus." Montgomery and Hill, in 1940, observed that both previously described lesions represented the same disease. He discussed the different lichen planus clinical and histopathological characteristics and ruled out the possibility that this lesion was one of its variants, as some scholars believed. Thus, this disease was described as "lichen sclerosus and atrophicus," but the term "atrophicus" was later withdrawn, since not all lichen sclerosus cases are atrophic [8, 5].

Lichen sclerosus lesions involving extragenital sites are reported only in a minority of cases [6], and oral cavity involvement is rare. To our knowledge, only 39 cases have been reported in the literature (PubMed, accessed on 10/17/2018) since its first description by Miller et al. [9], (**Table 1**). Most oral LS cases affect women with a mean age of 34 years and are usually localized lesions that are not associated with the presence of lesions in other anatomical locations.

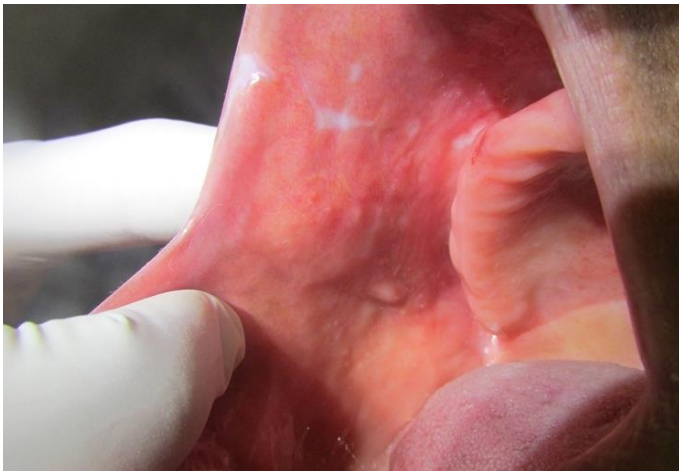

Figure 1. *Leukoplakic lesions in the right buccal mucosa, extending towards the labial commissure.*

Table 1 summarizes the characteristics of the oral LS cases reported in the literature, as well as the one described herein. This survey indicates a greater prevalence of LS in females and reveals that this condition may affect several oral mucosa locations, such as the buccal mucosa, tongue, hard palate, soft palate, lip vermilion, and gingiva. The lesions usually present as whitish diffuse or single plaques, of variable size, which may or may not be associated with pain or itching. In the case reported herein, the presence of several non-detachable whitish plaques was observed in the right buccal mucosa, extending to the proximity of the labial commissure and upper vestibule fundus. This clinical aspect is similar to what is commonly reported in the literature [1, 3].

Clinically, it may be difficult to distinguish LS from other oral white lesions, especially since this entity is quite uncommon in the oral cavity [3]. The differential diagnosis includes localized scleroderma, systemic scleroderma, leukoplakia or squamous cell carcinoma, lichen planus, vitiligo, and oral submucous fibrosis [1, 21]. Thus, a biopsy is fundamental for the correct diagnosis of oral LS.

Histopathological LS characteristics are typical and generally exclude the aforementioned conditions, although differences may be subtle [1, 3]. Oral LS exhibits six main important features: (1) hyperkeratosis, atrophy or focal degeneration of basal cells of the oral mucosa epithelium; (2) subepithelial cleft; (3) hyalinization of the superficial lamina propria; (4) a diffuse, irregular or band-like underlying mononuclear inflammatory infiltrate; (5) perivascular inflammation; and (6) telangiectatic blood vessels [3, 31]. Among these characteristics, the most striking are the hyalinization of the lamina propria and the underlying inflammatory mononuclear infiltrate. In the present case, all six morphological characteristics were present, leading to a definitive oral LS diagnosis.

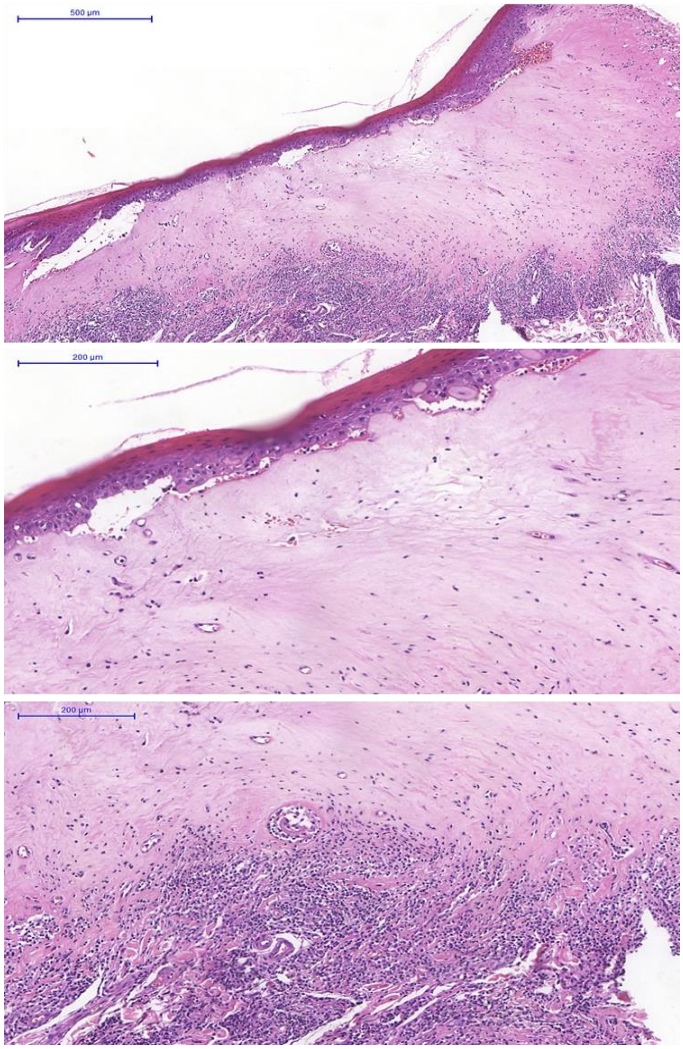

Figure 2. ***A)*** *General appearance of the lesion, presenting parakeratinized stratified squamous epithelium with the presence of a subepithelial cleft. Fibrous connective tissue exhibiting a hyalinized and homogeneous appearance and, in its deeper portions, the presence of an intense lymphocytic infiltrate, H&E.* ***B)*** *In more detail, the lamina propria presenting intense band hyalinization, H&E.* ***C)*** *Lymphocytic inflammatory infiltrate in the band underlying the hyalinization region of the fibrous connective tissue, H&E.*

The patient was referred to a dermatologist and gynecologist, revealing the absence of dermatological and anogenital lesions. This is important, as genital lesions may undergo malignant transformation, progressing to squamous cell carcinoma [3]. In the present case, because of the 8-year persistence and for aesthetic reasons, total surgical removal was performed, confirming the previous histopathological diagnosis. In turn, although the surgical approach has been adopted in some cases [1, 26-28]. However, reports indicate that observation may be employed in cases not associated with painful symptomatology [7, 25-27, 3]. In cases associated with symptomatology, topical corticosteroids may be used. Compared to systemic drugs, topical medications such as triamcinolone, betamethasone, and clobetasol have fewer side effects and have demonstrated better efficacy in the partial or complete resolution of the disease, making this treatment the first choice for symptomatic cases [3, 31]. Regarding asymptomatic cases, owing to the lack of studies in the literature there is still no consensus as to the best therapeutic approach. In the present case, after one year of follow-up, there are no clinical signs and recurrence of symptoms, but the patient remains under follow-up.

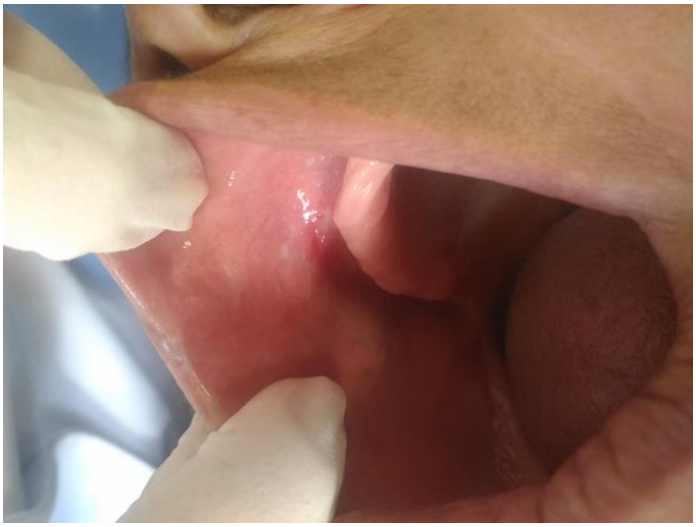

Figure 3. *Absence of lesion recurrence after 6 months of follow-up.*

Conclusion

In conclusion, reports in the literature emphasize that LS in the oral cavity is rare, and this diagnosis should be considered when non-detachable whitish plaques are observed in the oral cavity, as observed in our patient. The importance of the histological presence of subepithelial hyalinization and mononuclear inflammatory infiltrate in establishing a definitive LS diagnosis is highlighted.

Potential conflicts of interest

The authors declare no conflicts of interests.

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Table 1. *Summary of clinical cases reported in the literature*

	Authors	Sex/ Age	Location	Clinical aspects	Duration	Extraoral lesios	Treatment
1	Miller et al., 1957 [9]	F/48	Buccal Mucosa	White plaque. Asymptomatic	7 months	Yes	None
2	Ravits & Welsh 1957[10]	M/24	Buccal Mucosa, gingiva and hard palate	Multiples atrophic whitish-gray plaques. Asymptomatic.	4 months	NR	Vitamin A
		M /42	Buccal mucosa and palate	Multiples atrophic whitish-gray plaques. Asymptomatic.	NR	Yes	Vitamin A and D
3	Siar & Ng, 1984[11]	M/25	Buccal mucosa and lips	Multiple whitish plaques. Asymptomatic.	6 months	Yes	Corticosteroids
4	De Araújo et al., 1985 [12]	F/26	Upper gingiva and buccal sulcus	White plaque. Asymptomatic.	NR	NR	NR
5	Macleod et al 1991[13]	F/57	Língua and hard palate	Multiple atrophic white plaques with erosion areas	Several months	No	Corticosteroids
6	Schulten et al 1993 [14]	F/59	Labial mucosa and tongue	Multiple well-defined white plaques Size: 0.5, 1.5 and 0.2 in diameter. Asymptomatic	3 months	NR	None
		M/12	Lower lip vermilion	Well defined white plaque. Size: 0.8cm. Asymptomatic	9 months	No	Surgical excision
7	Brown et al, 1997 [15]	M/44	Soft palate	Slightly raised white lesion. Size: 1.0 9 1.5 cm. Single lesion. Asymptomatic	NR	No	Topical and intralesional corticosteroids
		M/18	Lower lip vermilion	Multiple whitish macules. Asymptomatic .	4 months	No	Topical and intralesional corticosteroids
8	Buajeeb et al, 1999 [16]	F/22	Mucobuccal fold, Buccal Mucosa and lower lip vermilion	White macular lesion. Size: 7.0 9 2.0 cm. Single lesion. Pain during tooth brushing. Tightness on opening the mouth	NR	No	Topical and intralesional corticosteroids
9	Jimenezet et al., 2002[17]	F/19	Upper lip frenulum, Buccal sulcus and upper gingiva	Well-defined whitish. Size: 1.0 cm in diameter. Mild discomfort	NR	NR	Intralesional corticosteroids
10	Kaur et al., 2002 [18]	M/16	Upper lip and Gingiva	White plaque. Size: 4,0 x 2,0cm. Mild discomfort	12 months	Yes	Intralesional corticosteroids
11	Jensen et al, 2002 [19]	F/10	Buccal mucosa and sulcus	Whitish plaque. Asymptomatic.	12 months	No	None

12	Rajlawat et al., 2004 [20]	F//14	Lower lip vermilion	Macular, ivory-white lesion. Size: 3.0 x 1.5 cm. Asymptomatic.	12 months	No	Intralesional corticosteroids
13	Mendonça et al., 2004 [21]	F/20	Lower lip vermilion	White macule. Size: 7.0 9 2.0 cm. Single lesion. Asymptomatic	NR	No	None
14	Kelly et al., 2006 [22]	F/10	Lower lip vermilion	Well-demarcated, atrophic white plaque with a central fissure. Size: 1.5 x 1.2 cm. Asymptomatic	2 years	No	Intralesional corticosteroids
15	Jimenezet et al., 2008[23]	F/31	Vestibular gingiva	White plaque Pain during tooth brushing.	2 years	No	Intralesional corticosteroids
16	Azevedo et al., 2009 [1]	M/19	Lower labial mucosa	White plaque. Asymptomatic.	8 months	No	None
		F/34	Upper lip vermilion and buccal sulcus	White plaque with ulcera área. Burning and pruritus.	3 months	No	Surgical excision
		F/11	Upper lip vermilion	White plaque. Asymptomatic	3 years	Yes	None
		F/38	Lower lip, lower lip vermilion and buccal mucosa	White plaque. Soft pain and pruritus.	20 days	Yes	Intralesional corticosteroids
		F/31	Lower labial mucosa	White plaque. Asymptomatic	6 months	Yes	Corticosteroids and Surgical excision
		F/28	Labial mucosa, buccal mucosa and tongue	White plaque. Mild discomfort.	7 months	No	None
17	Sherlin et al., 2010 [2]	M/60	Bilateral on the Buccal Mucosa and Gingiva	Multiples white patches with reddish areas. Size: 2.0 x 2.0 cm and 3.0x 2.0 cm. . Asymptomatic	6 months	Yes	None
18	Kim et al., 2010 [24]	F/7	Lower lip vermilion and alveolar mucosa	Well-demarcated, creamy white, atrophic plaque with sclerosis and telangiectasia. Size: 2.5x 1.5 cm. Asymptomatic	2 years	No	Topical corticosteroids

19	Liu et al., 2013 [7]	F/54	Tongue	Well-demarcated, atrophic, porcelain white plaque. Size: approximately 2.0 x 3.0 cm. Asymptomatic	10 days	No	Topical corticosteroids
		F/58	Tongue	White plaque. Size: approximately 2.5–3.0 cm. Asymptomatic	NR	No	Topical corticosteroids
20	George et al. 2014 [25]	M/20	Upper lip vermilion	Mutiple white lesions. Size: 1 cm. Asymptomatic	NR	NR	None
21	De Aquino et al., 2014 [26]	M/ 46	Upper lip frenulum and alveolar mucosa	Well-limited white patche with red areas. Size: 2.0 x 1.5 cm. Asymptomatic	NR	No	Surgical excision
22	Tupsakhare et al., 2016[27]	M/67	Soft palate	Well-demarcated whitish-gray plaque. Size: 0.8 x 0.8 cm. Asymptomatic	56 anos	No	Surgical excision
23	Marangon-Júnior et al. 2017 [3]	M/24	Upper lip vermilion	Well defined, ivory-white macules. Size: 3.5 x 2.0 cm and 0.5 cm in diameter. Multiple lesions. Mild, intermittent discomfort	2 months	NR	None
24	Kakko et al., 2018 [28]	F/28	Soft palate and gingiva	Multiple difuses white plaques	14 months	No	Surgical excision
		F/52	Soft palate and alveolar mucosa	Multiple white plaques with leucoplasic aspect	NR	No	NR
25	Matela; Hagström; Ruokonen, et al., 2018 [29]	F/70	Upper and lower attached gingiva	Porcelain-white, well-demarcated, non-scrapable, smooth lesions. Size: upper jaw gingiva: 4 x 1 cm; lower jaw: 3 x 0.5 cm. Asymptomatic	1 year	Yes	NR
26	Robledo-Sierra et al., 2018 [30]	F/65	Upper labial mucosa and right buccal mucosa	White macular lesion; burning sensation on the tongue.	9 months	Yes	Topical corticosteroids for symptoms
		F/68	Lower lip; soft palate	Discrete erythema and white striations over the surface of the lower lip, with features of atrophy; white, macular, and homogenous lesion.	2 years	Yes	NR

		M/13	Tongue and lower lip	Whitish lesion, resembling a fibrous scar or some form of atrophy (tongue); White macule (lower lip)	1 year	Yes	NR
27	Present case	F/ 57	Buccal mucosa	Multiple White plaques. Size: 1,0 x 2,0 cm and 0,5cm diameter. Asymptomatic	8 years	No	Surgical excision